CLINICAL PRESENTATION AND OUTCOMES OF PAEDIATRIC AUTOIMMUNE HEMOLYTIC ANEMIA – A RETROSPECTIVE ANALYSIS

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Autoimmune hemolytic anemia (AIHA) is a rare disease in childhood characterized by autoantibodies against erythrocyte membrane, causing excessive or uncompensated hemolysis and a clinical range from mild to severe life-threatening anemia.

We conducted a retrospective study of 18 AIHA patients treated at the University Children's Hospital Skopje between 2018 and 2022.

The mean age at diagnosis was 4.92 years, with 61% males and 39% females. Common manifestations included fever, jaundice, hepatomegaly or splenomegaly, with 61% exhibiting two or more features. The mean hemoglobin level at presentation was 63 g/L, necessitating red blood cell transfusion in all cases. Direct antiglobulin test (DAT) was negative in only one patient. Indirect antiglobulin test (IAT) was positive in 50%. Acute transient AIHA was observed in 16 patients, while 2 patients had chronic AIHA. Notably, both chronic patients were also diagnosed with Gilbert's syndrome. Primary AIHA was diagnosed in 44% of the cohort and all of them achieved remission after an average 4-week corticosteroid treatment. Secondary AIHA was identified in 56%, predominantly associated with infections, while one patient had Evans syndrome and one had autoimmune pancreatitis. Patients with chronic secondary AIHA received a combination of corticosteroids, immunoglobulins, rituximab and immunosuppressants. All other secondary AIHA patients responded well to corticosteroid monotherapy or a short course of immunoglobulins.

Secondary AIHA patients experienced a prolonged recovery compared to those with primary AIHA, who achieved complete remission. These findings highlight the importance of appropriate investigation and treatment strategies for AIHA.